



Case Report / Olgu Sunumu

An extremely rare lung tumor of a young adult: Primary synovial sarcoma

Genç bir yetişkinin çok nadir bir akciğer tümörü: Primer sinoviyal sarkom

Ali Özdil¹, Huriye Gülistan Bozdağ¹, Banu Yaman², Alpaslan Çakan¹, Ufuk Çağırıcı¹

¹Department of Thoracic Surgery, Ege University School of Medicine, İzmir, Turkey

²Department of Pathology, Ege University School of Medicine, İzmir, Turkey

ABSTRACT

Primary lung sarcoma is an extremely rare tumor accounting for less than 0.5% of all malignant lung tumors. Diagnosis is mostly established postoperatively. In this article, we present a 29-year-old male patient with a lobulated and well-marginated mass in the lower lobe of the right lung. A diagnosis of monophasic fibrous synovial sarcoma of the lung was established after right lower lobectomy. Resection margins were tumor-free. Postoperative positron-emission tomography/computed tomography did not show any other primary or metastatic focus. Despite the lack of a preoperative diagnosis, the advised treatment in such tumors is complete resection ensuring tumor-free surgical margins.

Keywords: Lung; primary; synovial sarcoma.

ÖZ

Primer akciğer sarkomu tüm malign akciğer tümörlerinin %0.5'inden daha azını oluşturan çok nadir bir tümördür. Tanı çoğunlukla ameliyat sonrası konulur. Bu çalışmada sağ akciğer alt lobunda lobüle ve düzgün kenarlı kitlesi olan 29 yaşında bir erkek hasta sunuldu. Sağ alt lobektomiden sonra akciğerin monofazik fibröz sinoviyal sarkomu tanısı konuldu. Rezeksiyon sınırlarında tümör yoktu. Ameliyat sonrası pozitron emisyon tomografi/bilgisayarlı tomografisinde başka bir primer veya metastatik odak görülmedi. Ameliyat öncesi tanı olmamasına rağmen, bu tümörlerde önerilen tedavi tümörsüz cerrahi sınır sağlayacak şekilde komplet rezeksiyondur.

Anahtar sözcükler: Akciğer; primer; sinoviyal sarkom.

Primary lung sarcoma is extremely rare and accounts for less than 0.5% of all malignant lung tumors.^[1] Its most common variants are leiomyosarcoma, malignant fibrous histiocytoma, and primary synovial sarcoma.^[2] These tumors may originate from the parenchyma, tracheobronchial tree or pulmonary artery and are described as mesenchymal tumors in the World Health Organization classification.^[3]

The preoperative diagnosis of synovial sarcomas is challenging since they have no radiological features. Thus, most diagnoses are made postoperatively. The recommended treatment for primary synovial sarcoma of the lung is surgical resection, if possible.^[4,5] In this article, we report a patient of primary synovial sarcoma of the lung who underwent curative surgical resection and was diagnosed postoperatively.

CASE REPORT

A 29-year-old male patient with complaint of right-sided chest pain was admitted to the hospital. A supradiaphragmatic lobulated opacity was detected on his chest X-ray. He was a non-smoker with no history of familial malignant diseases. Physical, clinical, and laboratory examinations were unremarkable. Chest computed tomography revealed a 115×83 mm, lobulated and well-marginated mass in the right lower lobe. There were no hilar or mediastinal lymphadenopathies (Figure 1). A written informed consent was obtained from the patient.

First, he underwent a right-sided videothoroscopic exploration which revealed a mass in the lower lobe that had not invaded the diaphragm or mediastinum, but invaded the visceral pleura with whitish tissue changes.

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Correspondence: Ali Özdil, MD, Ege Üniversitesi Tıp Fakültesi Göğüs Cerrahisi Anabilim Dalı, 35100 Bornova, İzmir, Turkey.

Tel: +90 232 - 390 49 19 e-mail: dr_aliozdil@yahoo.com

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Biopsy for frozen section was not taken because the resection of the lesion was not possible without lobectomy. Right lower lobectomy and mediastinal lymph node sampling from all lymph node stations via thoracotomy were performed (Figure 1). The postoperative course was uneventful and the patient was discharged on postoperative day seven.

The tumor was well-circumscribed, hemorrhagic and necrotic with dimensions of 90×52 mm macroscopically. Microscopically, it consisted of spindle-shaped cells with uniform appearance and small amounts of indistinct cytoplasm with ovoid dark-staining nuclei. Necrosis and increased mitotic activity were seen (12 mitoses per 10 high-power fields). Immunohistochemically, the tumor was stained with bcl-2 and TLE-1 diffusely. Cytokeratin AE1/AE3 was scattered positive. CD34, S100, and CD117 were negative. Ki-67 proliferation index was 12% (Figure 2). A diagnosis of monophasic fibrous synovial sarcoma was established with these investigations. Resection margins were tumor-free and mediastinal lymph nodes were reactive.

After histopathological diagnosis, the patient was reevaluated with positron-emission tomography/computed tomography (PET/CT) for another

sarcomatoid primary tumor and/or its metastases. No radiopathological finding was revealed. Adjuvant chemotherapy was planned for the patient by oncologists. The patient is under follow-up for 14 months without any recurrence.

DISCUSSION

Synovial sarcoma is a mesenchymal tumor accounting for 10% of all soft tissue tumors. It is an aggressive malignant neoplasm with a slight male preponderance and occurs most commonly in adolescents and young adults in soft tissues of extremities.^[6,7] It has also been observed in the head and neck, mediastinum, prostate, lung, and many other organs.^[5] Clinical, radiological, pathological, and immunohistochemical investigations are required to exclude other primary tumors and metastatic sarcoma.^[7] Our case was a young adult as reported in the literature in terms of the high-risk age group.

Primary synovial sarcoma of the lung is rare and there are few published series in the literature. Approximately 90 cases have been reported with an age range of 12-81 years.

Synovial cell sarcomas are classified as biphasic, monophasic fibrous, monophasic epithelial, and

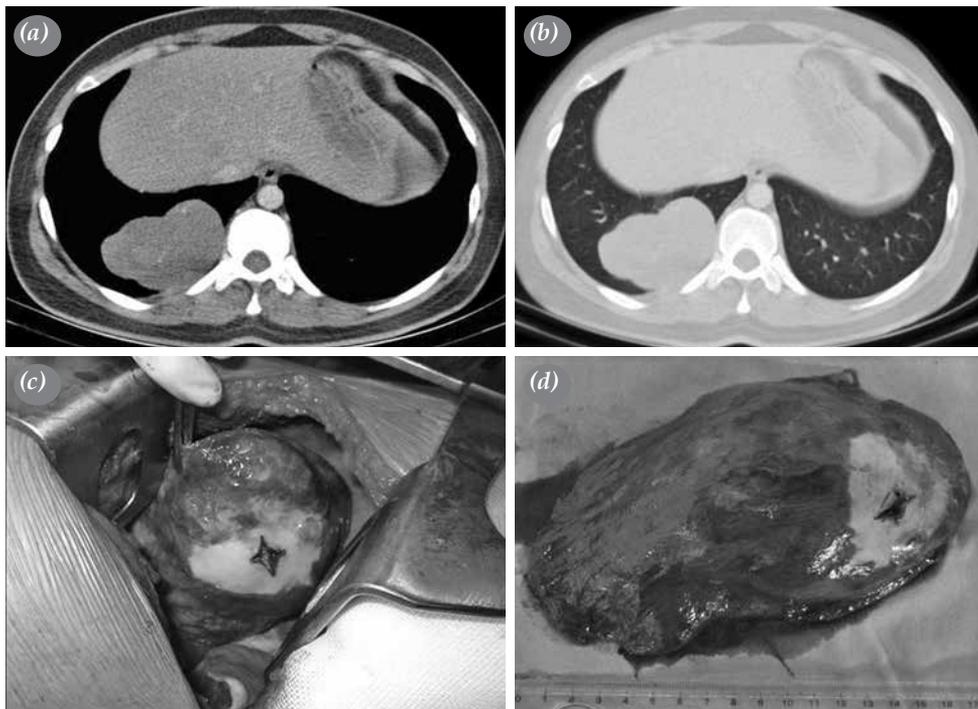


Figure 1. (a) Thorax computed tomography showing a large, lobulated, and well-marginated mass in right lower lobe of lung. (b) Intraoperative sight of tumor in right lower lobe and tumor in right lower lobectomy material.

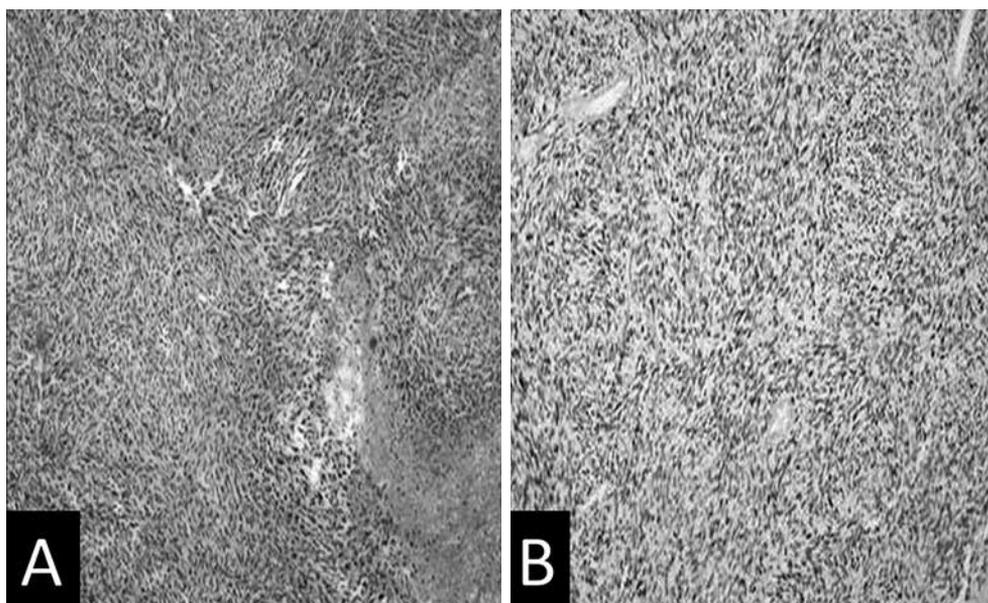


Figure 2. (a) Synovial sarcoma composed of spindle cells with round-ovoid nuclei (H-E×200). (b) Immunohistochemically bcl-2 positivity of tumor cells (×100).

poorly differentiated and may be confused with other spindle cell sarcomas.^[3] The differential diagnosis of spindle cell tumors may be difficult particularly in needle biopsies. Close differential diagnosis of synovial sarcoma includes fibrosarcoma, hemangiopericytoma, leiomyosarcoma, and other spindle cell tumors. Total excision of the mass is required to evaluate all the features of the tumor and study the immunohistochemical properties on a large panel; therefore, most diagnoses are established postoperatively. Before accepting the tumor as a primary lung sarcoma, extrathoracic areas should be examined in detail by radiologically. As mentioned above, we confirmed a diagnosis of monophasic fibrous synovial sarcoma by histopathological investigations after total excision and did not detect any extrathoracic area on PET/CT.

Complete resection with free surgical margin is the mainstay of treatment.^[1] The resection of nearby structures, if involved, must be included. There is no consensus or recommendation about the optimal surgical treatment of primary synovial sarcoma of lung.^[8] Although the role of adjuvant chemotherapy is unclear, it should be offered for avoidance of recurrence and prolongation of survival.^[3] The overall five-year survival rate is approximately 50%. Poor prognostic factors are age >20 years, tumor size >5 cm, positive margin of resection, mitotic activity >10 per 10 high-power fields, and the presence of

SYT-SSX1 variant.^[9] If localized recurrences are observed, resection should be performed with the aim of negative margins.^[1] We performed lobectomy to our case for the diagnosis and treatment of a large mass in the right lower lobe. The resection seemed to be curative because the surgical margins were negative and no lymph node metastasis was observed. Adjuvant chemotherapy was started due to the high-risk nature of the tumor and a long-term follow-up will be necessary for the result of treatment.

In conclusion, although primary synovial sarcoma of the lung is a rare diagnosis, it should be kept in mind in adolescents and young adults without history of any predisposing factor. Extrathoracic areas must be investigated for any other, particularly soft tissue, origin. Adjuvant chemotherapy should be offered for long-term survival after R₀ resection.

Declaration of conflicting interests

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